Case Report

Atypical Ischemic Cardiomyopathy after Resolution of a Giant Interventricular Septal Hematoma after Repair of Ventricular Septal Defect

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We present a 5-month-old girl who had interventricular septal hematoma (IVSH) after repair of ventricular septal defect. Intraoperative transesophageal echocardiogram showed that a giant hematoma occupied the interventricular septum immediately after coming off cardiopulmonary bypass. The patient was conservatively treated because hemodynamics was stable. Subsequently, the IVSH disappeared 3 weeks after the surgery. Follow-up cardiac catheterization 1 year after the operation showed normal coronary arteries including septal branches, but left ventricular function remained impaired. Transthoracic echocardiogram demonstrated thinning of the ventricular septum and hypokinetic motion of the septal wall. Myocardial perfusion scintigraphy and cardiac magnetic resonance demonstrated myocardial infarction in the mid-septal area consistent with the resolved IVSH. This suggested atypical ischemic cardiomyopathy. At the last follow-up (2 years after surgery), she has no symptoms of cardiac failure on oral administration of β-blocker, angiotensin-converting enzyme inhibitors, and diuretics for ventricular dysfunction. Patients with IVSH should be carefully followed even after hematoma disappeared.

Keywords: congenital heart surgery, interventricular septal hematoma, ischemic cardiomyopathy, ventricular septal defect

Introduction

Interventricular septal hematoma (IVSH) is an extremely rare complication associated with myocardial infarction, chest wall trauma or various cardiac procedures; they are responsible for disturbance and reperfusion of coronary blood flow to the septum.[1, 2)] Most of IVSH associated with congenital heart surgery are reported after repair of ventricular septal defect (VSD).[3)] IVSH occasionally results in life-threating events, such as ventricular systolic or diastolic dysfunction, arrhythmia, and ventricular outflow tract obstruction,[4)] depending on the size and the location of IVSH. Therefore, patients with IVSH should be treated according to the degree of hemodynamic compromise. Their outcomes in the intermediate term, nonetheless, have not been clarified yet, especially in pediatric patients associated with congenital heart disease, because information remains rather limited. Herein, we report a case of atypical ischemic cardiomyopathy following a giant IVSH conservatively resolved.

Case Report

A 5-month-old girl who weighed 4.7 kg failing to thrive was admitted to our hospital with a diagnosis of VSD. Transthoracic echocardiogram (TTE) revealed

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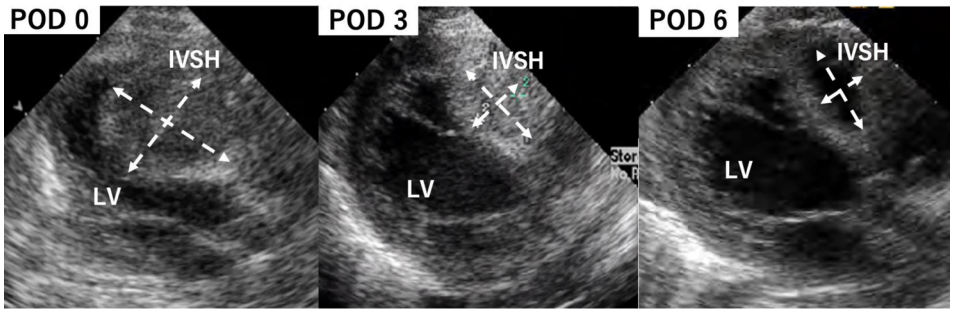

Fig. 1 Consecutive images of the hematoma at days 0, 3 and 6 after the operation showing spontaneous regression
IVSH, interventricular septal hematoma; LV, left ventricle; POD, postoperative day.

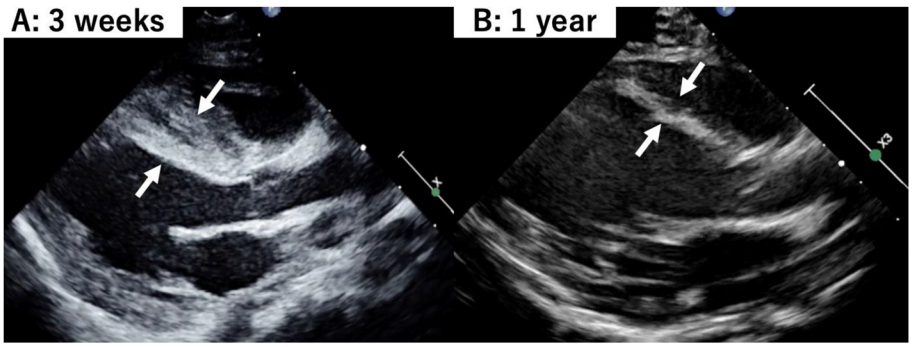

Fig. 2 (A) TTE shows a nearly normal septal thickness at discharge (white arrow). (B) TTE 1 year after surgery shows thinning of the ventricular septum (white arrow).
TTE, transthoracic echocardiography.

that the VSD was large and perimembranous of a trabecular type with posterior malaligned. No left ventricular asynergy noted. Preoperative TTE showed the left ventricular end-diastolic dimension and ejection fraction (LVEF) was 34.6 mm (Z-score; 5.94) and 62.7%, respectively. Preoperative cardiac catheterization showed that the ratio of pulmonary to systemic blood flow was 3.07, and the right and the left coronary arteries had normal origins without any stenosis. The VSD was surgically closed using an expanded polytetrafluoroethylene patch with interrupted mattress sutures through the right atrium. Coming off cardiopulmonary bypass was uneventful on intravenous dopamine (5 μg/kg/min) and milrinone (0.3 μg/kg/min). Intraoperative transesophageal echocardiography showed giant hypo-echogenic thickening of the interventricular septum. It was confirmed that the hypo-echogenic area did not expand. Hemodynamics remained stable. We decided to avoid further surgical revision.

Arriving at the intensive care unit, the patient frequently showed premature ventricular contraction (PVC). An electrocardiogram showed anteroseptal elevation of ST-segment, and the serum creatine kinase-MB had risen (254 U/L). TTE showed a huge mixed echoic mass inside the ventricular septum (23 × 21 mm) with hypokinetic ventricular septal motion, but no obstruction across either of the ventricular outflow tracts (Fig. 1). We chose conservative therapy without surgical re-intervention because the patient remained hemodynamically stable. PVC could be under control on β blocker. Serial TTEs showed a spontaneous and substantial decrease in the size of the IVSH (Fig. 1). The serum creatine kinase-MB gradually settled. When the patient was discharged 3 weeks after the surgery, TTE showed a nearly normal septal thickness (Fig. 2). LVEF was 31.8% because of hypokinetic and paradoxical septal wall motion as well as global systolic dysfunction. Therefore, we initiated oral administration of β-blocker, angiotensin-converting enzyme inhibitors, and diuretics.

During the follow-up period, TTE showed thinning of the ventricular septum, hypokinetic motion of the septal wall, and impaired function of the left ventricle (Fig. 2). Cardiac catheterization 1 year after the surgery showed normal coronary arteries including septal branches (Fig.

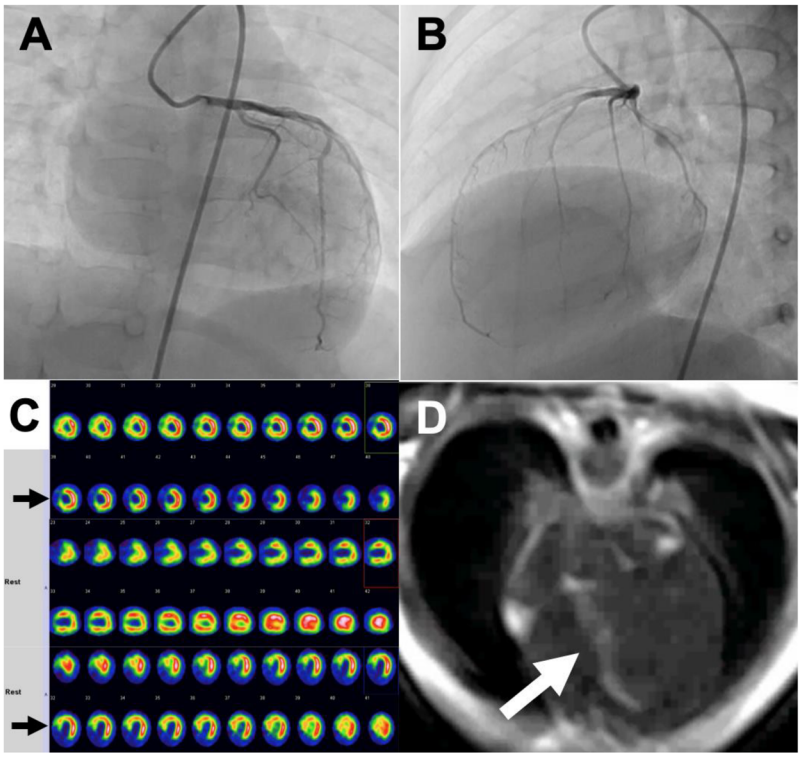

Fig. 3 Postoperative selective left coronary arterial angiography (A: frontal view, and B: lateral view). (C) Myocardial perfusion scintigraphy shows a perfusion defect (black arrow). (D) Cardiac magnetic resonance shows that the interventricular septum has late gadolinium enhancement (white arrow).

3A, 3B). Cardiac index 3.9. LVEF and LV end-diastolic volume was 35.6% and 39.4 mL (Z-score; 3.33), respectively. TTE also showed LVEF still low (41%). Myocardial perfusion scintigraphy using 99mTc-tetrofosmin at rest showed a perfusion defect in the mid-septal lesion (Fig. 3C). Cardiac magnetic resonance (CMR) showed late gadolinium enhancement in the interventricular septum (Fig. 3D). The patient is now 2 years old, on oral administration of β-blocker, angiotensin-converting enzyme inhibitors, and diuretics, without any heart failure symptoms.

Discussion

IVSH associated with congenital heart surgery was first described in 2005,[5] and to date, there have been 17 reported patients with IVSH after repair of VSD.[3, 4] Management strategies from these case reports are various, ranging from conservative to invasive. Approximately 70% of patients were conservatively managed, whereas the remaining had their hematoma surgically incised or needle punctured. There is no consensus regarding the management of IVSH associated with congenital heart surgery. Whether IVSH should be treated conservatively or invasively depends on the size and the location of the lesion; these factors could lead to hemodynamic compromise. The indication for surgical procedure would be rapid worsening of clinical conditions. On the other hand, aggressive surgical intervention is likely evitable, especially when patients' hemodynamics are stable.[3] Jegatheeswaran and colleagues have shown that patients with IVSH and hemodynamic incompetence are successfully treated using extracorporeal membrane oxygenation.[4]

The precise pathophysiology of IVSH remains unknown; it has been speculated that injury to the septal perforating artery (SPA) during placement of a VSD patch may be responsible for development of IVSH with signs of myocardial infarction.[5, 6] The SPA is at risk when suturing the patch to the anterior rim of the perimembranous VSD.[7] Hosseinpour and colleagues described that sutures for fixing a VSD patch should be carefully placed to prevent IVSH.[8]

Ischemic cardiomyopathy is typically defined as myocardial dysfunction that arises secondary to occlusive

or obstructive coronary arterial disease followed by degenerative changes in myocardial cells. In our patient, myocardial perfusion scintigraphy and CMR suggested a paucity of coronary perfusion and degeneration of myocardial cells in the ventricular septum consistent with resolved IVSH, which satisfied the diagnosis of "atypical" ischemic cardiomyopathy. The coronary angiography, nevertheless, showed a preserved SPA. This finding suggests a process as follow; injury to the SPA induced continuing bleeding to form an IVSH, the IVSH itself caused disruption of the microvascular circulation in the septal area resulting in an area of infarct, this evoked in turn thinning of the ventricular septum with impaired motion of the septal wall, and eventually lead to ischemic cardiomyopathy. Although a precise mechanism of pathophysiology remains unclear because histopathological assessment is missing, we assume that the IVSH caused myocardial ischemia and eventually irreversible myocardial stunning even after the space-occupying IVSH disappeared and even if the corresponding area contained normal myocardial cells and coronary arteries. There are only a few reports available regarding the mid-term outcomes of IVSH. Some reports mentioned an aneurysmal change of the left ventricle, akinesis of the septal wall, and lack of septal thickening, several years after surgery.[9)]

We described a case of atypical ischemic cardiomyopathy following resolution of a giant IVSH after repair of VSD. It is unclear whether or not surgical intervention for treating IVSH would prevent ischemic ventricular dysfunction after the hematoma disappeared, because those reports are quite limited. The optimal management remains undetermined and requires continuing evaluation of this lesion; IVSH is a rare lesion and its long-term outcome remains unknown. The patients with IVSH need careful and long-term follow up, even after remission of the hematoma, to scrutinize this injury which could lead to lethal arrhythmia, thrombus formation, or septal rupture. Furthermore, IVSH may cause not only these complications but also ventricular dysfunction, such as ischemic cardiomyopathy, in the longer term.

Conflict of Interest

All authors declare that they have no conflict of interest.

Ethical Standards

Informed consent for publication of this case report was obtained from the patient and the patient's guardians.

Author's Contribution

All authors (1) made substantial contributions to the study concept or the data analysis or interpretation; (2) drafted the manuscript or revised it critically for important intellectual content; (3) approved the final version of the manuscript to be published; and (4) agreed to be accountable for all aspects of the work.

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